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Congenital diaphragmatic hernia in children

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CONGENITAL DIAPHRAGMATIC HERNIA IN CHILDREN

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INTRODUCTION

Congenital diaphragmatic hernia is an uncommon condition which has created considerable interest because of the difficulty in clinical diagnosis (31). The diagnosis was previously too frequently "proved by necropsy," but now can easily be diagnosed by the use of X-ray and a curative surgical procedure can be applied (30).

The change in mortality shows the increased accuracy in diagnosis and early treatment. From 1912 to 1929, 81 cases were reported in patients less than ten years of age, of which only 25 cases were recognized during life (14). Prior to 1931, the medical literature reveals that about 75 per cent of the patients with congenital diaphragmatic hernia died before the age of one month. Potter (33), in 1952, said that only one infant at Chicago Lying-in Hospital with a diagnosis of congenital diaphragmatic hernia has survived more than six hours. In her experience, extrauterine respiration is rarely established because of secondary hypoplasia of the lung. In 1953, the mortality with surgical treatment was 25 per cent in patients less than one year of age (11).

The mortality rate for all types of diaphragmatic hernias from 1931 to 1940 was approximately 50

per cent. This has been improved to 10-15 per cent as shown by Zeller in 1950 (41). In 1945 Harrington (20) reported 430 cases of all types of diaphragmatic hernia in all ages which were treated surgically with a mortality of 4 per cent.

DEFINITIONS

Hernia: The protrusion of a loop or knuckle of an organ or tissue through an abnormal opening (Celsus) (10).

Diaphragm: Basically a dome-shaped musculofibrous partition between the thorax and the abdomen. It attaches to the xiphoid process anteriorly, the 7th to 12th costal cartilages laterally, and posteriorly to the bodies of the upper lumbar vertebrae by crus. The peripheral portion is muscular and the central portion consists of intertwining tendonous fibers (8).

Diaphragmatic Hernia: A protrusion of abdominal viscera through an anatomically weak area or through an abnormal opening or weak area in the diaphragm, the result of imperfect development, anatomical weakness or of trauma. A sac is a component part of a typical hernia, but the great majority of hernias through the diaphragm have no sac.

By common usage they are classed as false diaphragmatic hernias (24).

EMBRYOLOGY

Embryologically the diaphragm is formed by a fusion of mesodermal tissues of multiple origin. Development begins in the neck region at the level of the third cervical vertebra, from the 3rd and 4th myotomes which later form the ventral portion of the diaphragm. This Septum Transversum then migrates caudally carrying its nerve supply from the 3rd, 4th, and occasionally 5th cervical nerves to form the phrenic nerve. This anteromedial portion arising from the Septum Transversum finally divides the abdominal viscera from the heart in the primitive embryo.

During this caudal migration it fuses with mesodermal tissue from the dorsal mesentery forming the posteromedial portion. The lateral portions are thought to represent mesodermal tissue dissected from the body wall by the developing lungs.

The remaining bilateral defect, known as the pleuroperitoneal canals, is gradually obliterated by elements from the pleura above and the peritoneum below. The right canal usually closes first at about the eighth week, and the left canal about the tenth week. Migration

of muscle tissue into this pleuroperitoneal membrane completes the structure (1,8,32).

Separation of the two cavities is completed by about the third month of intrauterine life (1).

CLASSIFICATION

Diaphragmatic hernias have been classified embryologically, anatomically, and pathologically; but for clarity of diagnosis, the clinical classifications will be presented (13,20).

1. Non-traumatic Hernia
   a. Congenital
   b. Acquired after birth
2. Traumatic
   a. Direct injury
   b. Indirect injury
   c. Rupture of diaphragm due to inflammatory necrosis

Ricker (34), in 1954, categorized the congenital hernias in this fashion:

Congenital Diaphragmatic Hernia

1. Esophageal hiatus hernias
   a. Congenital short esophagus
   b. True hiatus hernias
2. Subcostosternal diaphragmatic hernias (Foramen of Morgagni)
3. Pleuroperitoneal hernias (Foramen of Bochdalek)

4. Eventration of the diaphragm

5. Congenital absence of the diaphragm

PATHOLOGIC EMBRYOLOGY

Congenital Short Esophagus

In early fetal life, if the stomach descends incompletely behind the Septum Transversum and does not reach the abdominal cavity before the lumbar portion of the diaphragm is completed, the esophagus may remain short and a portion of the stomach will remain in the thoracic cavity. Because the shortened esophagus enters the uppermost portion of the stomach, the normal relationship of cardia and fundus is lost. These are extremely rare hernias which can be demonstrated by X-ray examination with a barium or iodized oil swallow. Esophagoscopy will confirm the transition of esophageal to gastric mucosa well above the level of the diaphragm (34).

Esophageal Hiatus Hernia

Delay in descent of the stomach from the thoracic cavity to the abdominal cavity may keep the esophageal opening abnormally dilated during development of the diaphragm. Though the stomach eventually reaches its normal abdominal position and the esophagus attains normal length, this weakness in the esophageal hiatus will allow the

stomach and occasionally other viscera to herniate into the chest. These organs are always contained in a true hernial sac.

The hiatus hernias Ricker (34) has observed in infants have consisted of displacement of the upper portion of the stomach into the chest, including the fundus and cardia. The esophagus is of normal length but is buckled up in the mediastinum into a tortuous channel as seen on X-ray examination. These are not the common type of paraesophageal hernia which develop in older patients in which a knuckle of the fundus herniates alongside a normally placed esophagus.

This type of hernia rarely becomes symptomatic during infancy and childhood, but may be found incidentally on routine roentgenograms of the chest. The masses were then identified by fluoroscopy and barium meal.

Subcostosternal Hernias

Occurrence of herniation through the anterior defect in the diaphragm, known as the Foramen of Morgagni, has been explained as a failure of the central and lateral portions of the diaphragm to fuse. The confining sac usually present with such hernias suggests that the diaphragm has been completed by the layers of the pleural and peritoneal membranes, but that the growth of the muscle

between these two layers has been inadequate in this area, allowing intestines or liver to herniate into the thoracic cavity. These hernias are congenital but are rarely present at birth. They occur later due to increased abdominal pressure on a congenitally defective diaphragm (18).

Pleuroperitoneal Hernias

The separation of the common body cavity into the thoracic and abdominal compartments is accomplished by a series of orderly steps during the development of the embryo.

The Septum Transversum forms beneath the heart and grows backward to meet the dorsal mesentery of the foregut, completing the central portion of the diaphragm. A pleuroperitoneal fold then forms and gradually extends laterally and posteriorly to complete the division of the body cavities. The last portion to be bridged is posteriorly and is known as the pleuroperitoneal canal, or the Foramen of Bochdalek. Gradually the layers of pleura and peritoneum close this gap, and muscle fibers derived from cervical myotomes grow beneath the layers to form the final diaphragm. This is completed between the eighth and ninth week of fetal life. The left side of the diaphragm is completed later than the right,

possibly explaining the predominance of these hernias on the left.

During formation of the diaphragm the midgut lies out in the umbilical cord, where rapid elongation and development into its various components is taking place. The intestine returns to the abdominal cavity rather suddenly at about the tenth week of fetal life. Tardy closure of the diaphragm or too early return of the gut to the abdominal cavity may result in a portion of the newly returned intestine finding its way into the pleural cavity through the incompleted diaphragm. This plug of intestine probably prevents further closure of the diaphragmatic defect. Entry of the bowel into the pleural cavity before the pleuroperitoneal membrane has covered the defect results in a herniation without a confining sac. However, if the bowel pushes into the thorax after the pleural and peritoneal layers have formed but before muscular tissue has developed, the hernia will have a true sac (34).

Eventration of the Diaphragm

Eventration of the diaphragm seems to be a general stretching out of the diaphragm with an apparent relative diminution of the muscular elements about the periphery. The etiology may be a failure of the

muscular migration between the layers of pleura and peritoneum making up the diaphragm. If musculature were poorly formed, the increase in abdominal contents pushing upward and the development of normal intrathoracic negative pressure might result in a stretching out of the membranous diaphragm until it rises well into the chest (34). After repair of eventration the two sides of the diaphragm move synchronously, therefore ruling out the possibility of phrenic nerve paralysis which may present an identical clinical syndrome (29).

Congenital Absence of the Diaphragm

This is a very rare condition which may be partial or complete absence of the diaphragm. This is a misnomer in that the anterior portion of the diaphragm is usually present but posteriorly and laterally absent. In actuality, this is a pleuroperitoneal hernia with a very large diaphragmatic defect (18,20).

Associated Congenital Defects

Potter (33) states that a congenital diaphragmatic defect is one of the most common forms of defective development which occurs as an isolated disturbance in otherwise normal infants.

It is of interest to note that occasionally

other abnormalities occur with congenital diaphragmatic hernia. These are, in order of frequency:

1. Malrotation of the intestines (37)
2. Harelip (24,39)
3. Cleft palate (24,39)
4. Patent ductus arteriosus or foramen ovale (24,39)
5. Abnormal or imperfect development of the lung, liver, or mesentery relationships (24,39)
6. Abnormal cranial development (24,39)
7. Monstrosities of various types (24,39)

INCIDENCE

The incidence of diaphragmatic hernia as indicated roentgenographically in a 1954 survey in Delaware was one per cent of the over-all population. They also stated that about only 50 per cent of diaphragmatic hernias may be diagnosed accurately with X-ray (26).

In 1925, Hedblom (22) covered the age incidence of 37 cases with congenital diaphragmatic hernia. Of the 37 cases, 67.6 per cent were male and 32.4 per cent were female; also about 15 per cent of the hernias in this series possessed a hernial sac.

Tabulation of the findings of Hedblom (22) will be found on following page.

Age	Cases	
1-10	8	..... Less than 1 yr. - 3 Cases 1-5 years - 3 " 6-10 " - 2 "
11-20	12	
21-30	6	
31-40	4	
41-50	1	
51-60	2	
61-70	0	
71-80	1	
No stated age	3	
	37	

Authorities now agree on an over-all occurrence of a hernial sac to be about 10 per cent; also, the left diaphragmatic hernia to be more frequent than the right in a ratio of 4-5 left to 1 right (28,41).

The most common congenital diaphragmatic hernia is through the Foramen of Bochdalek; next, the esophageal hernias; and least common, herniation through the Foramen of Morgagni (16,17,19,28,36,37,41). It is of interest to note that in studies of Foramen of Bochdalek hernias the left-sided hernias are more frequent than right-sided, possibly due to the earlier closure of the right diaphragmatic defect, and the protection offered the right

diaphragm by the liver mass (28,37). In Foramen of Morgagni hernias, which are hernias through a congenital defect produced by increased abdominal pressures, the right-sided hernia occurs more frequently than the left-sided hernias (13,16).

DIAGNOSIS

Congenital diaphragmatic hernia occurs more often than was previously suspected, and it should be ruled out in all cases showing obscure chest and upper abdominal symptoms. The physical signs in the chest are often so confusing that the diagnosis may be missed if a roentgenogram is not taken (9). The diagnosis may be made in 59.5 per cent by clinical examination or with X-ray; 30 per cent are made at operation; 6.3 per cent at post mortem examination; and 4.3 per cent remains uncertain (22).

The primary symptoms of congenital diaphragmatic hernia in patients less than one year of age are cyanosis, dyspnea, and vomiting. These three cardinal symptoms are related to the systems affected: the heart and great vessels, the respiratory system, and the digestive system (41). If the patient is older than one year of age the cardinal symptoms are vomiting, pain, and colic (21). These symptoms are due to mechanical interference with

the function of the organs herniated in whole or in part, and to the impairment of respiration and circulation.

In congenital diaphragmatic hernias the symptoms are variable and inconsistent but are all related to these systems. Greenwald and Steiner (40) collected 36 cases of congenital diaphragmatic hernia and tabulated the systems as follows:

Cyanosis	24	cases
Peculiar weak cry	8	"
Feeble attempt at respiration	8	"
Failure to breathe, but satisfactory heartbeat	8	"
Dyspnea	7	"
Difficulty with nursing	4	"
Convulsive attempts at respiration .	2	"
Cough	2	"
Vomiting	1	"
Weight loss	1	"
	36	"

In observing these patients it has been noted in some cases that the symptoms may be ameliorated by holding the infant in an upright position (31). With this symptom variability, it should be remembered that any perplexing abdominal, respiratory, or cardiac signs and symptoms may indicate the presence of diaphragmatic hernia (1).

If the abdominal contents are not within the thoracic cavity at birth they may be sucked up into the thoracic cavity as soon as the infant begins to cry. Therefore, in many cases the trouble starts with the first cry after delivery. The more the baby cries, the more abdominal viscera are sucked into the thorax, and the greater becomes the respiratory and cardiac embarrassment. This results in death from suffocation in a matter of minutes or hours. Because this is coincident with birth, the condition is too often confused with other pathologic processes (3). In the 90 per cent of the cases not possessing a hernial sac there may occur extensive prolapse of the stomach, small intestine, colon, and spleen into the pleural cavity. This, then, allows collapse of the lung on the involved side and permits the heart and mediastinum to shift to the opposite side which produces compression or hypoplasia of the opposite lung (41).

The hernias that occur through structural defects in the diaphragm almost always produce clinical symptoms in infancy, and most of the infants who are born with this type of hernia die in the first few hours or days of life of cardiac or respiratory embarrassment resulting from the marked unilateral alteration in thoracic pressure at a time when the compensatory reserve

has not been developed to a sufficient degree to maintain function (17). The early cardinal signs of dyspnea, cyanosis, and vomiting, as well as any perplexing upper abdominal, respiratory, or cardiac symptoms and signs should bring the attending physician to consider the possibility of diaphragmatic hernia (1).

The physical examination done in an orderly fashion may show many variable findings which point to the presence of a congenital diaphragmatic hernia. On inspection the asymmetry of the chest with a unilateral lag during inspiration may be evident. There is frequently a rapid respiratory rate. The scaphoid abdomen has been aptly described by Orr and Neff (31) as "giving the appearance of unnatural emptiness." This emptiness is the result of the condition where the abdominal viscera have forfeited the rights of residence in the abdomen (1,40). Palpation may reveal the deviation of the trachea which may indicate or alert the examiner to an abnormal condition in the chest. The shift of the cardiac P.M.I. aids verification of an abnormal condition (25). Percussion of the chest is characteristically variable and the change in the findings at different times should point toward the consideration of diaphragmatic hernia. There may be a shift in cardiac dullness, hyperresonance over areas of unaffected lung, decreased

resonance over partially collapsed lung, flatness over solid herniated viscera, and tympanitic sounds over hollow herniated viscera. On auscultation bronchial breath sounds are heard over areas of compensatory emphysema. Absence of breath sounds results from lung compression by the herniated viscera. A systolic murmur may occur if there is a shift in the position of the heart or great vessels. A pathognomonic sign for diaphragmatic hernia is the presence of borborygmi or intestinal rushes in the chest (25).

In eventrations, about half the patients are symptom-free and the symptoms and signs that occur are related to the respiratory, circulatory, or alimentary systems as in diaphragmatic hernias. There may be acute respiratory distress in the neonatal period, or multiple gastrointestinal complaints in the adults (29). The physical signs are inconsistent except for an increased respiratory excursion of the ribs on inspiration on the affected side. This has been called Hoover's or Korn's Sign which results from the absence of the inhibiting effect of the diaphragmatic excursion (4).

COMPLICATIONS

The most frequently encountered complications in congenital diaphragmatic hernia are acute respiratory

failure and acute intestinal obstruction. The respiratory failure occurs early in life, but if compensated for by the patient's physiology, acute intestinal obstruction becomes a definite hazard (24). The mortality increases with obstruction of the alimentary tract to 60 per cent, whereas the mortality without obstruction is equivalent to any other abdominal operation. This emphasizes the importance of early diagnosis and definitive surgical treatment. The size of the defect is related to the possibility of obstruction in that obstruction is more likely to occur if the defect is small than if it is large (5,21).

Frequently associated with congenital diaphragmatic hernia is a failure of rotation of the colon, and occasionally the small bowel may be enveloped in a congenital peritoneal fold which constitutes an internal hernia. In 58 per cent of congenital diaphragmatic hernia there is malformation of the mesentery of both the small and large intestine more usually called "malrotation of the bowel" (17).

There is little relationship of the viscera displaced into the chest and the mortality, except when the small bowel is herniated, whence the mortality is definitely increased (21).

Strangulation of a congenital diaphragmatic hernia

may occur but is very rare (35). Carter and Giuseffi (6) reported four cases of strangulation of a congenital diaphragmatic hernia, and Wolfson and Goldman (39) reported one case of congenital strangulated diaphragmatic hernia of the liver. Strangulation is defined as the arrest of circulation due to compression of the herniated tissue.

ROENTGENOLOGY

The increasing use of X-ray and its technical advances have added greatly to the accurate and more rapid diagnosis of congenital diaphragmatic hernias. The important observations seen on the X-ray film will indicate the size of the hernial mass, the state of compression of the lungs, and the position of the heart and mediastinum. Seldom can the size of the diaphragmatic defect be accurately predicted (15).

Both fluoroscopic examination and roentgenogram films may be of help. The characteristic findings in the chest are as follows: (a) poor lung expansion on the affected side; (b) mediastinal shift to the unaffected side; (c) heart shift to the unaffected side; (d) the presence of abdominal viscera on the affected side continuous with those present in the abdomen. If a hernial sac is present it tends to limit the herniated viscera to the lower lung field, but if the sac is absent, the

viscera tend to ascend higher in the thorax and may even reach the apical region (28). There are many important details to observe in less obvious cases which may require the use of thin barium meal to bring out the specific herniated viscera. The promiscuous use of barium should be avoided because of the possibility of vomiting followed by aspiration. The barium is very irritating to the respiratory tract and will complicate the patient's treatment and course of convalescence (12).

An additional diagnostic aid is the use of pneumoperitoneum. The air injected may outline the hernial mass on X-ray if a sac is present, and if the sac is absent the air may be seen at the apex of the chest on X-ray. If carefully done, the pneumoperitoneum may avoid the hazard and inconvenience of a diagnostic thoracotomy. The injection of 250-300 cc. of air into the peritoneal cavity may be done with minimal danger of air embolism, but should only be done by a trained individual (7).

HERNIATED VISCERA

The order of frequency of the herniated contents, according to Greenwald and Steiner (14), is stomach, transverse colon, omentum, small intestine, liver, pancreas, and kidney. Hedblom (24), in a review of 1,003 cases, showed that the stomach was present in the

hernial contents in 69 per cent of the cases. In 71 per cent of the cases the colon and small intestine were herniated with other organs. Also of significance is the fact that only 56 of these 1,003 were right-sided hernias in which 11 had only liver tissue involved in the hernia. Harrington (18,20), in his series of 430 cases of diaphragmatic hernia which came to operation, has shown the type of hernia has a definite relation to the viscera involved. Of the eight cases of the Foramen of Morgagni type, the colon was involved most frequently, next the omentum, and lastly the ileo-cecal coil. Only rarely was the stomach involved in this type of hernia. The frequency with which the stomach occurs in diaphragmatic hernias is due to the author's including the esophageal hiatus hernias in adults in their series.

DIFFERENTIAL DIAGNOSIS

The differential diagnosis of congenital diaphragmatic hernia in the newborn has been presented by Wyatt (40). The differential diagnosis is as follows:

1. Intracranial hemorrhage.--The newborn is usually hypertonic or spastic and convulsions frequently occur. Irregular respiration and cyanosis may simulate congenital diaphragmatic hernia. The nystagmus and pupillary findings indicate cerebral injury. If

vomiting is present, then X-ray should be used.

2. Laryngeal obstruction.--Chevalier Jackson described laryngeal dyspnea as an indrawing at the supersternal notch, around the clavicles, and at the epigastrum. This should easily be differentiated from congenital diaphragmatic hernia in most cases.
3. Atelectasis.--Cyanosis is present and rapid respiration may be present. Dullness and bronchial breathing can usually be elicited.
4. Massive collapse of the lung.--This condition is usually due to a plug of mucous in the main bronchus. According to Tow, the onset is sudden, with cyanosis, labored breathing, dullness, bronchial, diminished or absent breath sounds on the injured side. X-ray shows the heart to be shifted toward the involved side.
5. Congenital pneumothorax.--Cyanosis and dyspnea are sudden in onset. The heart is pushed to the opposite side. X-ray is of distinct value.
6. Congenital heart disease.--Dyspnea, orthopnea, cyanosis may be present or absent, rapid respiratory rate. Auscultation and X-ray aid in diagnosis.

In infants, older children, and adults the differential diagnosis should include the following items:

1. Eventration.--J. Dewey Bisgard (4) collected 183 cases from the literature and found that eventration is very

rarely symptomatic in infancy. The etiology is obscure, but probably most cases are congenital in origin. The symptoms are respiratory, circulatory, and alimentary as in congenital diaphragmatic hernias.

2. Pleurisy.--This may be with or without an exudate, and can usually be differentiated from congenital diaphragmatic hernia by the clinical history.
3. Pulmonary tuberculosis.--A family history of the disease or a close social contact may be elicited. X-ray is of prime importance in early diagnosis.
4. Primary intrathoracic region tumor.--Early diagnosis only with the use of X-ray. Some tumors of the lower lobes of the lungs may be mistaken for small diaphragmatic hernias, and vice versa.
5. Primary gall bladder disease.--The symptoms of gall bladder disease in early childhood may be difficult to differentiate from an asymptomatic diaphragmatic hernia (25).

TREATMENT

The early treatment of patients with congenital diaphragmatic hernia, in order to improve function of the respiratory and alimentary systems, may include turning the baby on the side of the hernia to allow the the heart and mediastinum to fall away from the lung on

the opposite side, affording better aeration (15). The infant may be held in an upright position to allow the abdominal viscera which has herniated into the chest to gravitate into the abdomen and thereby relieve some of the partial obstructive or alimentary distresses (31).

The policy of delaying surgery until the child is older and stronger has resulted in greater mortality than surgery done before the infant is 48 hours old, and provides the added advantage of having the intestines deflated. The collapsed alimentary tract facilitates reduction of the hernia and makes the abdominal closure much easier (28).

As previously discussed, the treatment is surgical because of the high mortality and frequent complications if treated conservatively.

Preoperative Preparation

It is important to have the patient in the best possible condition for surgery without excessive delay. This is accomplished by proper hydration, alimentary deflation, and respiratory aeration (27,28). Hydration is accomplished by giving subcutaneous, intravenous, or rectal solutions as indicated (2,27,28). Deflation is accomplished by the use of gastric suction and a rectal tube. Adequate aeration of the lungs and oxygenation of

the tissues is most satisfactorily done by placing the patient in an oxygen tent with a high oxygen content and humidity. The high oxygen concentration serves two purposes: (a) making the patient's breathing less labored, and (b) by helping to keep the intestine deflated.

Anesthesia

Ochsner (30) advocates the use of an endotracheal tube and a closed system positive pressure technique because of the greater ability to control the patient and provide an adequate supply of oxygen. Ochsner also reported one case of absence of the anterior mediastinum associated with congenital diaphragmatic hernia in which bilateral collapse of the lungs would have occurred had the patient not have been intubated. The anesthetic agent of choice in all reports is cyclopropane because it provides a maximum oxygen content, is easily handled in the closed positive pressure system, and gives good muscle relaxation in the patient at the time of operation. Also of great importance is the presence of a trained anesthesiologist to handle the precise needs of the patient.

Phrenic Crush

A preliminary phrenic crush, done through a small superclavicular incision on the affected side (27,28),

just prior to the operation for repair of the hernia, is not essential but may be desirable. It is easily accomplished by the use of a hemostat and the effects of diaphragmatic paralysis on the affected side will last from five to six weeks. It will allow the diaphragm to be at rest during the operation and thereby make the closure of the defect easier. The diaphragm will be high, due to the loss of phrenic innervation and will therefore increase the room in the abdomen for the hernial contents after reduction. There will be less tension, the suture lines allowing better healing. This procedure is primarily used in adults rather than in infants.

Surgery

The thoracic approach was previously considered good because it enabled the surgeon to close the defect in the diaphragm with greater ease. A profound disadvantage was encountered in trying to reduce the herniated viscera into the abdominal cavity because the cavity had become too small.

The abdominal approach has been found to be more advantageous than the thoracic route because the reduction of the hernia is easily accomplished and the defect may be closed satisfactorily. There are no adhesions formed in the thorax which hold the abdominal viscera and, therefore, reduction is easy if done in an orderly

fashion. The negative pressure in the thorax holds the abdominal viscera securely until the suction is relieved by passing a catheter through the diaphragmatic defect. The viscera should be removed from the thorax in an orderly fashion. On the left side the stomach should be reduced first, then the small intestine, cecum, ascending colon, splenic flexure, and lastly, the spleen. On the right side the intestines should be removed first in the same order as above and lastly, the liver should be reduced (28).

The frequency with which malrotation occurs and the possibility of intestinal obstruction, strangulation, or volvulus, and the occasional need to do a bowel resection make the abdominal approach definitely superior (37). The use of the thoracic approach initially may require an extension of the incision into the abdomen on a second abdominal approach to complete the procedure of repair if malrotation or obstruction is present (17).

Following reduction of the abdominal viscera, the diaphragmatic defect is closed with mattress sutures after denuding the margins of the defect. The peritoneal layer is then sutured separately on the underside of the diaphragm. The pleural cavity is then obliterated by withdrawing all the air possible to aid expansion of the lung.

If the abdominal viscera cannot be placed in the abdominal cavity with ease a two-stage closure of the abdominal wall may be done. This is described by Ladd and Gross (28) as first closing only the skin and subcutaneout tissues after undercutting the skin. The second stage is performed five to six days later before the cutaneous sutures have cut through the skin and after allowing the abdominal wall to become stretched out to accommodate the viscera which had previously been in the chest. The second stage closure should be a layered closure of the abdominal wall. This two-stage closure is superior to prolonged efforts of the usual layered closure because it is less shocking to the patient. It also avoids placing undue strain on diaphragmatic suture line and it avoids pressure embarrassment on the respiratory, circulatory, and alimentary systems.

It has been stated that the abdominal approach has a higher postoperative mortality than the thoracic route. The use of abdominal approach for cases which are obstructed and for diagnostic laparotomies accounts for the greater mortality. For operations on diaphragmatic hernias which are not complicated by obstruction, the mortality is equal to or less than the mortality in cases done by the thoracic route (23).

Postoperative Care

The essential postoperative care is described by

Ladd and Gross (27). Blood transfusions are given for blood replacement and also to combat the hemorrhagic tendency of the newborn infant. An oxygen tent or an incubator with mist with an oxygen concentration of 90-95 per cent should be used to make breathing less labored, keep the intestine deflated, and remove rapidly any room air which might have been trapped in the pleural cavity. This high oxygen concentration should not be used in premature infants because of the possibility of producing retrolental fibroplasia, and should be carefully used in other full term newborn infants because of the irritating nature of high oxygen concentrations on the lung tissue. The maintenance of an adequate fluid balance is important for the first few postoperative days. This may be either oral or parenteral as necessary for the patient. The caloric requirements of the patient for the first few postoperative days do not have to be fulfilled.

Recurrence

The recurrence of diaphragmatic hernia following surgery in 378 cases surveyed by Hedblom (22) is about five per cent. Recurrence is primarily due to incomplete closure of the defect or excessive tension on the suture lines.

Surgical Mortality

The surgical mortality has been and will still be further decreased by the use of X-ray for early diagnosis of congenital diaphragmatic hernias. The improved anesthesia, better agents, and better methods of administration by trained anesthesiologists is important. The patient must receive adequate preoperative preparation and excellent postoperative care. The improved surgical techniques and recent advances for correct handling of the patients have been stressed by Hartzell (21). These are: (a) decompression of the alimentary tract and preoperative gastric lavage, (b) use of the phrenic crush, (c) aid in reducing the hernia by use of a tube to break the thoracic suction, (d) the breaking or cutting of ribs to aid in repair of large defects, and (e) aspiration of the pneumothorax to increase postoperative lung expansion thereby decreasing dyspnea, cyanosis, and respiratory effort.

CAUSE OF DEATH

The common cause of death in the newborn and infants is primarily respiratory or cardiac. If the patient compensates for the cardio-respiratory insult, he may then become obstructed, strangulated, and may then die of electrolyte imbalance or by perforation with

peritonitis (14,21,23).

In the postoperative patient death usually occurs within 48 hours and is primarily due to respiratory or cardiac failure, with partial atelectasis of the lung (17).

CASE PRESENTATIONS

Case No. 1

Children's Memorial Hospital #25,102
24-hour-old white male

Presenting Picture: Dyspnea
Cyanosis
Rapid forced respiration with sternal retraction
Left thorax splinted with respiration
Right basilar rales
Left lung hyperresonant to percussion
Heart shifted to right

X-ray - Chest Film: Heart displaced to the right
Left lung only 70 per cent expanded
Entire intestine in the left thorax

Surgery: Endotracheal--Cyclopropane plus oxygen
Abdominal approach
Defect of Foramen of Bochdalek (3 cm. dia.)
Sac present
Hernial contents--entire small bowel
--proximal and middle and distal colon
Complications--postoperative pneumothorax
--occasional postoperative vomiting
Result--good

Other Defects: None

Discussion

This case presents the typical picture of Foramen of Bochdalek congenital diaphragmatic hernia with the exception that a hernial sac was present.

Case No. 2

University of Nebraska Hospital #121,256
5-day-old white male

Presenting Picture:
- Weak cry at birth
- Respirations not vigorous for first five minutes
- Rapid respiratory rate
- Cyanosis
- Dyspnea
- Vomiting and regurgitation after meals
- Improved respiration, decreased vomiting in upright position
- Flat abdomen
- Liver down to umbilicus in right mid clavicular line
- P.M.I. to right of sternum
- Right tracheal shift
- Right shift of heart and mediastinum
- Borborygmi in left chest

X-ray:
- Left diaphragm high
- Overdistended right lung and left apical lobe
- Poorly defined left heart border

Surgery:
- Endotracheal--Cyclopropane and oxygen
  --Ether - Nitrous Oxide and oxygen
- Abdominal approach
- Defect Foramen of Bochdalek
- Sac absent
- Hernial contents--Stomach
  Small intestine
  Colon
  Spleen
  Left lobe of Liver

Complications--
Jejunal fistula present (Resection and anastomosis)
2 Dehiscence episodes
2 Stage closure of abdomen
Final Result--good

Other Defects: None

Discussion:

This case presents the classical Triad of symptoms and characteristic physical findings. The jejunal defect could not be explained at the time of operation.

The patient had a stormy course with two episodes of wound dehiscence due to tension on the abdominal sutureline when the abdominal contents were reduced from the hernia.

The final layered closure was accomplished and the infant dismissed in good condition.

Case No. 3

Children's Memorial Hospital #13,656
3-month-old white male

Presenting Picture: Bluish pallor
Rapid shallow respiratory motions
Frequent upper respiratory infections and "chest colds"
Subnormal appetite
Decreased breath sounds over Posterior mid portion of right chest
Left bronchial breath sounds
Tachycardia--Heart rate - 125
--Aortic 2nd sound was greater than the Pulmonic 2nd sound

X-ray:	Rounded cystic mass of Soft tissue density 4½ cm. in diameter occupying the lower half of the Right Anterior mediastinum. Heart displaced to Left with Prominent apex of Left Ventricle.
Impression:	Dermoid cyst Bronchiogenic cyst Enterogenous cyst
Surgery:	Endotracheal-Ether and oxygen Initial Thoracic Approach of the Exploratory Thoracotomy Secondary Abdominal Approach Defect - Foramen of Bochdalek Sac present Hernial contents - Liver Complications: Postoperative shock and respiratory distress Abdominal distension Shallow Respirations Cyanosis Result--Patient expired less than 6 hours postoperative
Other Defects:	Deformed Right Ear Ptosis of Right eyelid Prominent parietal bosses Pupils react slowly to light Mongolism

Discussion:

This case was misdiagnosed by X-ray, and was difficult due to the hernial tissue being solid liver. The hernia was small and confined with a sac and the symptoms were quite pointed to the respiratory tract.

The patient was subjected to both a thoracic and abdominal approach. In this case it might have been repaired through the thoracic approach without the loss of the patient.

The general condition of the patient with various deformities and the diagnosis of Mongolism probably had considerable influence on the final demise. No autopsy was obtained to determine if there were any defects in the heart or in the central nervous system.

Case No. 4

Children's Memorial Hospital #19,443
2½-year-old white female

Presenting Picture: Heavy breathing and panting for two months
Respiratory Rate--Sitting - 42
Supine - 60
Increased AP diameter of chest with Pigeon breast deformity
Decreased breath sounds in left chest
Dullness over posterior left lung field below 3rd rib
Flatness of Anterior right chest to right mid-clavicular line (heart)
P.M.I. 2 cm. to right of sternum
Grade I systolic murmur at right sternal border
Active bowel rushes over entire lower left lung field
Trachea deviates to right

X-ray - Chest Film: Trachea and heart shifted to the right
Consolidation of middle left lung field
Atelectasis of lower left lung field due to compression
Gas loops of large and small bowel in left lower chest
Lateral Chest Film--
Suggests Foramen of Bochdalek hernia
Gastrointestinal Series--
Small bowel and right colon in left chest
Splenic flexure and descending colon in normal position

Surgery:
- Endotracheal--Ether and oxygen
- Abdominal approach
- Defect -- left Foramen of Bochdalek (6 x 3½ cm.)
- Sac absent
- Hernial contents--
  - Jejunum
  - Ileum
  - Appendix
  - Caecum
  - Ascending and transverse colon
- Complications--None
- Layered primary closure
- Result--good

Other Defects:
- Partial malrotation of bowel
- Pigeon breast deformity

Discussion:

The physical findings on clinical examination and X-ray were the basis for diagnosis of congenital Diaphragmatic hernia in this patient.

The results were good.

Case No. 5

University of Nebraska Hospital #120,221
4-year-old white male

Presenting Picture:
- Frequent Upper Respiratory Infections since birth
- Slight underdevelopment in height and weight for chronological age
- Trachea deviated to right
- Dullness and tympanitic sounds to percussion over lower left lung field
- Decreased breath sounds over left lung
- P.M.I. in the 5th intercostal space, 2 cm. to the left of the mid-sternal line
- Heart shifted to right

X-ray - Chest Film: Right tracheal and mediastinal shift
Heart shifted to right
Atelectasis of left lung base
Small bowel in left chest
Gastrointestinal Series--
Small bowel in left chest
Appendix and caecum in apex of left chest

Surgery: Endotracheal--Ether--Cyclopropane and oxygen
Thoraco-abdominal approach
Defect--Foramen of Bochdalek
Sac absent
Hernial contents--
Entire small intestine distal to ligament of Treitz
Caecum and appendix
Ascending and transverse colon
Complications--None
Layered abdominal primary closure
Results--Excellent

Other Defects: Right inguinal hernia

Discussion:

This patient presented with a minimal amount of respiratory symptoms but had quite definite physical signs which were confirmed on X-ray.

The patient had lived four years, but has not developed as he should have, probably due to the frequent upper respiratory infections and the difficulty in digestion of food due to the mal-placed alimentary tract.

Case No. 6

Children's Memorial Hospital #24,640
6½-year-old white female

Presenting Picture:	Vague "Stomach Ache" since age three years--intermittent without relation to eating, activity, etc. Occasional brown vomitus Abdominal pain and vomiting every day for past 3 months Right diaphragm seemed higher than normal to percussion
X-ray -Fluoroscopy:	Paradoxical motion of diaphragm on coughing. (Right diaphragm elevates and left diaphragm descends.) Gastrointestinal Series-- Stomach low in abdomen Entire small intestine and proximal colon were between the liver and diaphragm Descending colon travels from right upper quadrant to rectum directly
Surgery:	Endotracheal--Ether-Nitrous Oxide and oxygen Abdominal approach Defect--Foramen of Bochdalek (5 x 7½ cm.) Sac present Hernial contents-- Entire small intestine and proximal colon Small right lobe of the liver Complications--None Layered abdominal primary closure Results--good
Other Defects:	Agenesis of right lobe of liver Enlarged left lobe of liver "Malposition" of bowel (Not true Mal-rotation)

Discussion:

This patient presented with the three cardinal complaints for congenital diaphragmatic hernia in patients over one year of age; i.e., pain, vomiting,

and colic.

The hernia was on the right side and possessed a sac which confined the hernial contents. This case was interesting because of the enlarged left lobe of the liver and the agenesis of the right lobe of the liver.

SUMMARY AND CONCLUSIONS

1. Congenital diaphragmatic hernia is a rare condition which has been changed from a fatal condition to one with a good survival by early diagnosis and immediate treatment.
2. The normal and pathologic embryology is presented to explain the present accepted theory for production of the various defects in the diaphragm.
3. Congenital diaphragmatic hernia is one of the most common forms of defective development which occurs as an isolated disturbance in otherwise normal infants.
4. The incidence of all types of diaphragmatic hernia is about one per cent in the general population.
5. Consideration of congenital diaphragmatic hernia is the most important step toward early diagnosis.
6. Cardinal symptoms of congenital diaphragmatic hernia are related to the circulatory, respiratory, and alimentary systems.
7. X-ray offers the best and most accurate aid for early diagnosis of congenital diaphragmatic hernia.

8. The most frequent complications are acute respiratory failure and acute intestinal obstruction.
9. Treatment of choice is early surgery which, if delayed, results in an increased mortality.
10. The mortality is definitely increased if the small intestine is involved in the hernial mass.
11. Surgical mortality has been decreased by immediate pre-operative preparation, careful anesthesia administered by a trained anesthesiologist, improved surgical technique, and excellent post-operative care.
12. Six cases are presented which came to operation at Children's Memorial Hospital and University of Nebraska Hospital from 1952 through 1955.

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